CHILDREN WITH DEVELOPMENTAL DISABILITIES AS CANDIDATES FOR COCHLEAR IMPLANTATION

Capstone Project

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By

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ABSTRACT

In recent years, the number of children with multiple disabilities and severe to profound hearing loss are receiving cochlear implants at an increasing rate (Kim et al. 2008). Preliminary research has established anecdotal data and small outcome case studies for this population. Although the literature has yet to evolve into conclusive findings and no standards of care for the candidacy of children with additional disabilities currently exist, the gains in clinical experience and professionals' confidence have led to more implant centers across the country beginning to consider and pursue this type of intervention for children with multiple disabilities. Interdisciplinary teams are an effective way to deliver services to the child and their family. These teams benefit professionals by helping them familiarize with child development and expertise of other team members in an effort to identify potential risk factors in their patients. These identifications can lead to appropriate and timely referrals to other health care providers. This Capstone advocates that with the right research and interdisciplinary teamwork, children with multiple disabilities can be appropriate candidates for cochlear implants.

DEDICATION

This capstone is dedicated to my loving husband, who has never stopped believing in me since the day when we first met. And to my parents, who have loved and encouraged me since the very beginning.

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TABLE OF CONTENTS

Page

Abstract……ii

Dedication……...iii

Acknowledgements……iv

Vita……v

List of Tables……vii

Chapters

1: Introduction……1

2: FDA Pediatric Candidacy……6

3: Interdisciplinary Cochlear Implant Teams……10

4: Special Considerations for Candidacy……26

5: Postoperative Outcomes……39

6: Conclusion……45

LIST OF TABLES

Table Page

1 Definition of Disability Categories…….3

2 Major and minor cochlear implant criteria…….9

CHAPTER ONE

Introduction

For every 1,000 live births, between 1 and 4 infants in the United States are identified with congenital hearing loss (Bass-Ringdahl, Ringdahl & Boelter, 2010; Kenna et al., 2007). Congenital hearing loss is defining as present and detectable using appropriate tests at or very soon after birth (Davis & Davis, 2011). Within this population, approximately 30 to 40 percent of children will have a co-existing developmental disability (Gallaudet Research Institute, 2003; Berrettini et al., 2008; Fortnum & Davis, 1997; Wiley, Jahnke, Meinzen-Derr, & Choo, 2005). This group of children is more difficult to define than those born with hearing loss alone; the manner in which multiple conditions coexist and the manner in how they are expressed are unique for every child (Diefendorf et al., 2011). In an attempt to categorize these children in a more straightforward fashion, the Individuals with Disabilities Education Act (IDEA, 2004) currently states that a child who has one or more physical, cognitive, communication, social or emotional, or adaptive development disabilities may be placed in just one category of developmental delay (Kreisman & John, 2011).

Although the law provides that these children can only be categorized by one disability, these children struggle with diverse associated disabilities. Specific types of developmental disabilities are linked as co-existing with a hearing loss in children with

multiple issues (Diefendorf, 2003; Roush, Holcomb, Roush, & Escolar, 2004). Gallaudet Research Institute (GRI, 2003) examined which developmental conditions were most prevalent in addition to a hearing loss. They found that co-existing disabilities were highest amongst those with a learning disability, cognitive disability, visual impairment, and cerebral palsy, respectively. Wiley et al. (2005) conducted a retrospective review on 35 children with multiple disabilities who received a cochlear implant. The authors revealed that hearing loss was linked to categories of visual impairment, motor disability/Cerebral Palsy, cognitive disability, learning disorder, behavioral disorder, or language disorder. If a child had more than one disability among this list in addition to hearing loss, they were listed as "additional disability". These definitions are summarized in Table 1 (Wiley et al., 2005). Learning disabilities are outside the scope of this Capstone as literature reveals too many definitions representing various problems and underlying causes for its diagnosis (Hendriksen et al., 2007).

Table 1 Definition of disability categories

Visual Impairment	Visual deficiency that cannot be corrected with corrective lenses or procedures to at least 20/70 or children who have a visual field of less than 20
Mild motor disabilities	Referred or enrolled in OT or PT but not meeting diagnostic criteria for cerebral palsy
Cerebral Palsy	Confirmed exam and documentation by a neurologist or developmental pediatrician
Cognitive disabilities	Borderline or lower non-verbal IQ (75 or lower) and delayed social-adaptive skills or developmental quotient (DQ) less than 70 in children under 6 years of age
Specific learning disability	Considered after the child has completed a multi-factorial evaluation and exhibits a discrepancy of academic performance from an average or above average non-verbal IQ
Behavioral disorders	Behavior disorders diagnosed by a physician including attention deficit disorder (ADD), oppositional defiant disorder (ODD), depression, and bipolar disorder
Language disorders	Diagnosed by stringent criteria following a comprehensive multi-factored evaluation; included autism, epileptiform aphasia, and receptive and expressive language disorder
>1 Additional disability	Having hearing loss and two or more additional disabilities
	Note: From "Perceived qualitative benefits of cochlear implants in children with multi-handicaps", by S. Wiley, M. Jahnke, J. Meinzen-Derr, and D. Choo, 2005, *International Journal of Pediatric Otorhinolaryngology*, 69, p. 793.

Fortnum, Marshall and Summerfield (2002) conducted an epidemiological study for 17,160 children born in the United Kingdom (UK) with a "permanent hearing loss" between 1980 and 1995. These children met the criteria by having a bilateral hearing loss of > 40 dB HL including conductive pathologies, since these children also require long term management. All participants were placed in categories for either moderate (i.e. 41-70 dB HL), severe (i.e. 71-95 dB HL) or profound (i.e. > 95 dB HL) degree of hearing loss. The authors found significant differences between all three groups. Particularly, children with moderate losses were more likely than those with more severe losses to have unknown causes or "unspecified aetiology" for their hearing loss. The children with severe losses were more likely than those with profound impairments to have perinatal complications (e.g. hypoxic ischemia, asphyxia) that were the cause of hearing loss. And

children with profound loss were more likely to have genetic causes (e.g. Down syndrome, Pendred syndrome, Usher syndrome) than those with lesser degrees.

Severe to profound hearing loss may be due to pre-natal, perinatal, or post-natal factors and in at least 50% of cases genetic factors are thought to play a part. A minority of children will have deafness as part of a well-recognized syndrome, such as Pendred or Usher that are important for healthcare providers to distinguish. A severe to profound hearing loss is fundamental criteria to establish when determining if a child should be selected as a cochlear implant candidate. During the candidacy assessment, the identification or specific etiology causing of deafness is particularly important as it may influence counseling about expectations, prognosis or the timing of the surgical procedure (Corina & Singleton, 2009; Daneshi & Hassanzadeh, 2007).

This Capstone systematically reviews current literature on children with multiple disabilities involving a severe to profound hearing loss, whose families are seeking the option to have their child receive a cochlear implant. The main purpose of this Capstone is to offer audiologists insight on the extremely high levels of care necessary for reviewing the implant candidacy of children and their families. Through the diligent use of an interdisciplinary team, this service for children with multiple disabilities can be achieved. Chapter 2 summarizes the United States Food and Drug Administration (FDA) regulations interdisciplinary team experts work within to help multiple disability children receive cochlear implants. Chapter 3 introduces the use and benefits of the unique nature of the interdisciplinary team. Chapter 4 identifies specific team criteria to determine candidacy for cochlear implants. Chapter 5 illustrates specific successful outcomes of

appropriate candidate children with multiple disabilities that benefited from interdisciplinary team expertise. Overall, this Capstone stresses the importance of providing quality service through a large team of specialists representing a variety of healthcare and educational fields. Their individual expertise will help evaluate and consider the complexities of candidacy, whose ultimate decisions and recommendations are voiced in a “single consultation” for the family.

CHAPTER TWO

FDA Pediatric Candidacy

Cochlear implant candidacy criteria for children and adults are regulated by the Food and Drug Administration (FDA, 2010). The FDA approved the use of cochlear implants in children as young as 12 months from evidence-based research demonstrating that prelingual deaf children are capable of developing speech, language and auditory skills (Cheng, Grant & Niparko, 1999; McConcey et al., 2004; Miyamoto, Houston, Kirk, Perdew, & Svirsky, 2003) and that the most effective strategy for normal language development is appropriate intervention of hearing loss by 6 months, whether through hearing aids or cochlear implants (Appuzzo & Yoshinaga-Itano, 1998; Yoshinaga-Itano, 2003; Yoshinaga-Itano, Sedey, Coulter, & Mehl, 1998). Further, the FDA outlines guidelines for appropriate pediatric candidacy selection. These guidelines include: a bilateral severe to profound hearing loss (e.g. > 70 dB HL), a physiologically intact cochlea and surviving auditory nerve fibers (i.e. spiral-ganglion cells), delays in developmental, social and communication milestones, parents with reasonable expectations concerning the outcomes of implant use, and minimal or no benefit from appropriately fit hearing aids. Lack of benefit in older children can be quantified by achieving a less than 30% open-set word recognition score on standardized tests including the Multi- syllabic Lexical Neighborhood Test (MLNT) or the Lexical Neighborhood Test (Kirk, Pisoni & Osberger, 1995). Parents of children younger than 4

years old must complete the Infant Toddler Meaningful Identification Scale (IT-MAIS Robbins, Renshaw, & Berry., 1991), a questionnaire that was originally developed for cochlear implant candidacy and allows parents to observe and record changes in their child's auditory behavior through hearing aid use.

Many implant facilities contest that additional areas for a child candidate should be considered including psychosocial functioning, language use and educational placement, as demonstrated by the inclusion of these criteria in assessment (Hayman & Kirk, 2005). In an attempt to organize the information collected during the candidacy process, Hellman et al. (1991) from the Manhattan Eye and Ear Institute established the Children's Implant Profile (ChIP) addressing factors that most contribute to success of implantation. There were originally 11 factors that have since evolved over twenty years of research including for example duration of deafness, mode of communication and nonverbal cognition level. All factors are rated on a 3-part scale for "level of concern" of a) none b) some or c) major. The Children's Hospital of Philadelphia (CHOP) has developed their own version of ChIP for evaluating every pediatric candidate, which is listed in Table 2.

The final decision on whether a child should receive a cochlear implant is ultimately determined by a team of professionals, which minimally includes a pediatric audiologist, speech-language pathologist, and otologic surgeon. The cochlear implant (CI) team must determine if the child's developmental and functional communication needs are best supported by a cochlear implant or hearing aid based on the FDA guidelines and their professional experience and expertise. If a child does not meet FDA guidelines, but the

team deems a cochlear implant to be medically necessary, then pending approval by the 3rd party payer (insurer), the team may move forward with what is known as ‘‘off-label’’ cochlear implantation (FDA, 2010). In this event, the FDA expects the team to maintain detailed records of the child’s progress and outcome with the cochlear implant. An “off-label” decision currently incorporates all candidates who have multiple disabilities since this population is not generally considered through FDA guidelines; however, this fact does not nor should not preclude any teams from making the appropriate recommendation.

Although any “off label” use in a child requires team input, this may be particularly true for children who are have multiple disabilities. The team that helps identify the appropriate pediatric candidates for cochlear implants fits within a larger interdisciplinary team that work with children with developmental disabilities. Team specialists work simultaneously on determining etiology and its underlying functional effects that are limiting the outcomes of development. This team approach is discussed in Chapter 3. The unique situations involving additional criteria necessary for this candidacy population is discussed in Chapter 4.

Table 2 Major and Minor Categories of the CHOP ChIP

Major Category	Minor Category
Chronological age	
Duration of deafness	Medical Radiological
Otolaryngology	
Other disabilities	
Audiology	Test reliability
	Hearing aid use
	Hearing aid benefit
Speech pathology	Auditory training
	Formal language use/comprehension
	Use of voice to communicate
	Desire to communicate
Social work	Family structure and support
	Child's behavior
	Expectations (parents)
	Expectations (child)
Education	Current educational placement
	Future educational placement (transition)
	Access to auditory-oral services
	Educational staff CI training
	Ability of parent to participate in educational process

CI-cochlear implant; CHOP- The Children's Hospital of Philadelphia; ChIP- Children's Implant Profile.
Note: From "Cochlear implant candidacy in children with autism" by C.D. Hayman and K. H. Franck, 2005, *Seminars in Hearing*, 26, p. 218.

CHAPTER THREE

Interdisciplinary Cochlear Implant Teams

To help provide comprehensive hearing healthcare for a child diagnosed with multiple disabilities, families should inquire about working with an interdisciplinary cochlear implant (CI) team. No two teams are alike, especially when working with a candidacy population with developmental delays in addition to severe to profound hearing loss. The cochlear implant process for these children will be referred colloquially as a "special case" (Filipo, Bosco, Mancini, & Ballantyne, 2004). The term implies team members are facing situations that are complicated beyond that of severe to profound hearing loss.

The different situations that surround a "special case" at any point in time have led implant facilities to establish their own criteria, which reflect a scattering of team philosophies regarding candidacy requirements of which children in this population. Some teams may adopt a more liberal attitude for recommending an implant which includes Children's Hospital of Philadelphia (CHOP) and Massachusetts Eye and Ear Institute (Mass Eye and Ear); others may place a greater emphasis on team input, weighting professional opinion more significantly than family counseling and considerations. Some teams, for example, may hold the philosophy that every family must have a thorough understanding of genetics influencing a case before making implant

decisions. Teams will stress this fundamental view by encouraging all parents to consider a genetic evaluation as part of the candidacy process. A differing team philosophy may feel it is best to steer clear of making a point of discussion regarding the value of genetic testing, since uncertainties exist about the moral or ethical beliefs held by a family. That is, some facilities may believe it prudent for teams to wait to engage a discussion about gene testing until it has been made clear by the family that they have an interest in pursuing the possibility. Although each team will have their own philosophy, the key factor is that a solid framework of specialists needs to work together to offer prime resources and counsel for the family and child with multiple issues to appropriately treat hearing loss and improve developmental and educational outcomes in these candidates.

System of Organization

The cochlear implant team must establish a cohesive, balanced and unified front through a system of organization for the interdisciplinary team of professionals. There are three major aspects to consider when implementing the ideal "special case" team. (1) Establishing an interdisciplinary team, as opposed to a multidisciplinary team, (2) focusing on directing care that is family centered, as opposed to professionally centered delivery service, and (3) incorporating suggested group models of a medical, educational and cultural perspective. When creating a team, the system of organization is important, and the involved professionals should consider these factors to arrange an effective team to help their unique child candidate. The type of framework a team constructs for providing healthcare management is important to discuss, because a chosen approach to

organization, no matter how subtle the differences seem, can create substantial differences in how a team functions and their potential outcomes offered to the family and child (Paul & Whitelaw, 2011).

First, the CI Team should be an interdisciplinary one as opposed to a multidisciplinary one. A multidisciplinary versus an interdisciplinary team approach can be difficult for people to differentiate, which has led many using of the two systems as interchangeable in their meaning (Paul & Whitelaw, 2011). This can be understood from an outsider perspective, as both approaches are incorporating a number of different professionals to represent the family and the coordination of care and management are "patient-centered" or "total patient" (Massachusetts Eye & Ear, 2009). However, this distinction is important; a CI team should emphasize an interdisciplinary approach.

Historically, teams used a multidisciplinary approach. Where the team functioned on a well-organized structural basis but whose conceptual framework portrayed power relationships between team specialists (Cass, Price, Reilly, Wisebeach, McConachie 1998) and between specialists and their patients (Decker, 1997). Power relationships create "role conflict", which has been described as overlapping responsibilities and "preconceptions that professionals have their own role on the team." (p. 3, Orchard, Curran & Kabene, 2005). These perceptions of the multidisciplinary team create a system of hierarchy (Cass et al., 1998), which restricts genuine considerations of another team member's opinion and perspective, specifically in assessing and recommending a child to receive a cochlear implant. More so, the delivery of intervention services via multidisciplinary approach inevitably creates power imbalances. These imbalances can

cause major conflicts among team members and can result in costing a hospital or implant facility more money from all the energy expended from frustrations and stubborn attitudes within the team (Orchard et al., 2005). Power imbalances are not only reflected on a professional-professional basis but are also apparent between the families. Orchard et al (2005) specifically noted that with this team approach, a child is viewed as the "patient" and parents are generally excluded from both the planning and decision-making process in respects to how to manage their child.

An interdisciplinary team on the other hand is promoting a true collaboration of effort between specialists, where high quality of care is not possible without professionals adopting a fundamental shift in attitude regarding for example the values they hold for providing care and the patterns of socialization displayed to the family and other professionals (Orchard et al., 2005). This shift brings different disciplines together, moving from isolation to promoting a balance of responsibilities, knowledge and skills (Jessup, 2005; Paul & Whitelaw, 2011). With the healthcare community adopting this approach on a more frequent basis, the goal-setting process and outcomes have become much improved versus if professions were working separately (Paul & Whitelaw, 2011), which will inevitably create delays from struggling with power conflicts (Orchard et al., 2005).

In order to maintain the cochlear implant philosophy that is brought forth by the team, an interdisciplinary approach demonstrates a consistent ability to communicate, brainstorm, adapt, and act in accordance with the team's philosophy, as each individual is interested in achieving a common set of goals. Goal setting is made on a case-by-case

basis, depending on family preferences and specific needs relating to the individual child (Choi & Pak, 2006). While on a special case implant team, it is important to remember that having many different fields of expertise may make it easy for miscommunication to occur such as one professional offering services or advice that is conflicting with a previously established goal. To avoid this, a special case team needs an interdisciplinary approach to ensure individual and collective responsibilities (Cass et al., 1998) that are shared equally among each professional.

Second, in accordance with an interdisciplinary approach that focuses on balance of responsibilities and practice (Cass et al, 1998) the choices in facilitating care and delivery of service is no longer mandated by one professional, which in past was the team physician. Rather, the delivery of service has shifted into family-centered practice with parents directing services and becoming the leading members of the interdisciplinary team for their child (Paul & White, 2011); the interdisciplinary CI team should be based on a family-centered model of care. The shift to family-centered care has helped to avoid delivery of service that has potential for creating professional bias. Family-centered model provides unbiased information regarding communication and educational decisions. The shift towards making parents the leaders of the team helps successful voice and implements change within the social and political environment (Paul & Whitelaw, 2011). This effective shift to a family-centered model is mandated through IDEA (2004) in early hearing loss detection and intervention services (EHDI) including Family-Guided Routine-Based Approach. EHDI programs are implemented to meet the needs of the family and child as a full package, including screening, diagnosis, early intervention and family support (Wang & Engler, 2011).

Thirdly, the team model (e.g., medical home, educational, or cultural) infuses knowledge into the team approach that operates as an interdisciplinary, family-centered system of organization. For example, the option of a special case team working within the medical home model is helpful for keeping a well-organized system of communication, and management (AAP, 2006). Specifically, reflecting accessible, coordinated, continuous, and family-centered care (Liptak et al., 2006). The family and child are the core team representatives, reiterating that the family is actively participating in discussions and always the ones to make the final decisions on whether to implant their child (Patel, Pratt, & Patel, 2008).

Members of the "Special Case" Interdisciplinary Team

Before management strategies are considered for the child, professionals on a "special case" team must have high levels of expertise to address underlying physiologic and functional concerns that are influencing the presenting delays in the child. Multiple developmental disabilities involving multiple organs and systems are frequently observed in children with concomitant hearing loss. Because of a multi-system involvement, it is critical that individual knowledge is demonstrated and can help for distinguishing the appropriate cochlear implant candidate. The hearing loss is often secondary in nature including for example craniofacial abnormalities in children diagnosed with Stickler, Cerebral Palsy (Zanchetta, Resende, Bentlin, Rugulo, & Trindade, 2010), CHARGE (Edwards, Kileny & van Riper, 2002), and Down syndrome. The assessment and input from each specialist must collectively lead to understanding the etiology of the hearing loss and concomitant disabilities and limitations for the individual child.

The following is a list of professionals on a "special case" team who are capable of addressing such issues.

- Pediatric Audiologist- The pediatric audiologist clearly has an important role on the interdisciplinary team working with cochlear implants and children. A pediatric audiologist on a "special case" must be able to demonstrate a general frame of knowledge on the "common" syndromic conditions known to co-occur with a hearing loss (Diefendorf, 2003; Maddel & Flexer, 2008; Hood & Keats, 2011). The audiologist must demonstrate a high level of flexibility while administering and collecting behavioral responses for a child with multiple disabilities, including visual response audiometry (VRA) and conditioned play audiometry (CPA). The testing situations will never be the same from one day to the next because you are dealing with a child with cognitive delay and multiple disabilities that may need various adjustments to testing procedures. Additionally, throughout the cochlear implant candidacy period and on-going care of the child, other audiologists may join or provide inputs to the special case team. For example, the educational audiologist will be helpful in communicating audiologic information with the speech-language pathologist, teacher of the Deaf, early interventionist and others within the educational system. During team meetings the educational audiologist is critical for emphasizing interrelationship between the child's degree of hearing loss, auditory skill development, rate of spoken language, and level of cognition (Chute & Nevins, 2011). During a candidacy evaluation, audiologists must be able to communicate to non-speech and hearing

professionals the potential outcomes of benefit and limitations if a child was to receive a cochlear implant.

- Physical Therapists- Functional delays that are due to physical and motor-related disabilities can co-exist with a concomitant hearing loss in this population, such as those with cerebral palsy. The physical therapist on a "special case" team offers expertise related to the degree to which a physical deficit is restricting the child's current functional abilities. That is, specialists serve in answering questions behind which physical and motor-related factors are limiting the child's abilities to utilize a full range of motion with their limbs, display typical outcomes in gait, have adequate head, neck and trunk control, structural posture and stability, show capability of manipulating small objects, and overall dexterity levels that are parallel to their peers of the same age who are typical and demonstrating functional milestones within an appropriate time frame (Costigan & Light, 2011; Hong & Zolli, 2005). The therapists will assist the audiologic responsibilities of obtaining thresholds of hearing while testing in the soundbooth by maintaining the child's postural stability. For example, in order to get accurate behavior responses from the VRA, the child requires the appropriate level of head, neck and trunk control (Madell & Flexer, 2008). In these situations, the physical therapist is accommodating a procedural modification that will help the audiologist gather reliable behavioral outcomes; it also helps in highlighting the child's strengths and skills that they are capable achieving once the appropriate accommodations are made.

- Developmental/Behavioral Pediatricians (DBP)- These physicians are essential for special case candidacy, as their expertise will help in coordinating the diagnosis and management for children including hearing loss, visual impairment, cognitive and motor disabilities, and other issues, such as epileptic seizures (Accardo & Shapiro, 2006). These specialists are the physicians that the child and family most frequently visit and are most likely to develop the closest bond with the family and child (Boreman, Thomasgard, Fernandez, & Coury, 2007).
- Psychologist- The psychologist's role is to address behavior and cognition. That is, without having the expertise that helps to tease out the "why" involving children with co-existing communication, behavioral and social interaction delays the interrelated effects from a hearing loss will never be completely understood. Thus, the psychologist has a critical role for predicting growth of auditory skills and potential in benefiting from a cochlear implant. For example, the Diagnostic and Statistical Manual of Mental Disorders, 4th edition (DSM-IV; Frances, Pincus & First, 2000) is the "gold standard" for assessing and diagnosing conditions including autism and a broad spectrum of cognitive behavior disabilities. However, tests that rely on spoken language are invalid measures to test on children with a concomitant hearing loss, since language is clearly delayed from significant auditory deprivation. A psychologist on a "special case" team must be familiar with finding alternative testing measures such as *Battelle Developmental Screening Inventory, 2nd Edition* (BDIS-II; Glascoe & Byrne, 1993, Matson, Hess, Sipes, & Horovitz, 2010) and the *Leiter International Performance Scale- Revised* **(**LIPS-R; Leiter, 2002). These tests assess nonverbal ability for cognition as

opposed to IQ or language based tests. In addition to functional measures, the psychologist on a "special case" team must have a "trained eye" for observing both apparent and subtle nuances in behavior for early identification of autism (Vernon & Rhodes, 2009), where characteristics may not manifest until the child gets older. For example if the diagnosis of autism has been established in the child, the team psychologist can collaborate by sending the audiologist outcome reports identifying maladaptive behavioral traits of the child before the audiologic candidacy evaluation. The audiologist may gain information on how a certain time frame exists before the child "zones out" from hypo-responsiveness; another child with autism spectrum disorder may demonstrate hyper-responsiveness to sensory stimuli, which produces extremely negative reactions to a loud sound or tactile contact whether it being from another person touching the child or the sensation from probe inserts or a cochlear implant processor (Hayman & Franck, 2005).

- Otologist/Otolaryngologist- The otologist or otolaryngologist on a "special case" team must have clinical experience in evaluating and treating children with craniofacial disorders including the external and middle ear components, oral region, jaw and mandibular portion, and the size and shape of the skull. Some or all of the dysmorphic traits are presenting in children with a diagnosis such as Down syndrome or Treacher-Collins syndrome (Hood & Keats, 2011). The expertise of the professional must reflect an ability to minimize complications of one or more physical features that are contributing to secondary effects of hearing loss. Otologists need to offer input on whether the dysmorphic assessment

displays contraindications for the child's ability to physically retain the weight of an external implant processor. The abnormal properties of skull shape, size or thickness may not allow a cochlear implant device to maintain proper stabilization and adherence to the side of the head.

- Ophthalmologist- Many forms of syndromic hearing loss are accompanied by visual defects including coloboma of the eye (Edwards et al., 2002), ocular lesions and maculopathy (Hood &Keats, 2011), and progressive blindness via retinosis pigmentosis (RP) disease. Vision loss is sometimes not present at birth, which is common in many forms of Usher syndrome; late-onset symptoms have historically made the confirmation of diagnosis difficult at birth. This has led to many children receiving a misdiagnosis of nonsyndromic hearing loss until a later determination of vision loss (Hood & Keats, 2011). It is therefore important for the team ophthalmologist to be monitoring these children from birth. They should track the status of an identified eye disorder and/or catch problems as soon as revealed. This will help for determining prognosis of having optimal amounts of auditory stimulation for communication through spoken language because they will not have visual modes of communication.
- Nephrologist- Children with multiple disabilities are at higher risk for displaying health problems relating to the renal system. Problems involving kidney dysfunction are common with certain syndromic hearing losses including children diagnosed with Alport syndrome, Branchio-Oto-Renal (BOR) syndrome and Distal Renal Tubular Acidosis (dTRA) (Hood & Keats, 2011). Nephrologists who have experience working with children are needed for examining whether the

kidneys are contributing to the surrounding issues of the child. Screening and monitoring kidney function are critical for children who have been identified with a specific condition or who are been placed at risk for manifesting future symptoms of renal disorder. Specifically, frequent urinalysis may reveal microscopic hematuria, or blood in the urine of the child, which is the most common and earliest sign in children born with Alport syndrome. Alport syndrome is a progressive disorder involving both eventual kidney failure and bilateral profound hearing loss (Keats, Berlin & Gregory, 2006; Nance, 2003). If symptoms of the disease are caught early, kidney treatment such as renal transplantation can be maximized in the outcomes for the child (Kashtan, 2006). The nephrologist would be able to determine if, and when, the candidate child was medically capable of undergoing surgery for the cochlear implant.

- Neurologist- The neurologist may address structural and functional issues of the brain. The administration of clinical examinations to establish neurological status will help in defining disorders of cranial nerves III, IV and V (i.e. controlling eye movements), VII (i.e. facial sensation and movement). These problems in the cranial nerves may cause auditory neuropathy spectrum disorder (ANSD) and those in the NICU are at increased risk for presenting this disorder (Rance, Dowell, Rickards, Beer, & Clark, 1999)
- Social Workers- Special counseling skills are required to uphold the family dynamics whose parenting skills, coping methods, financial circumstance, and cultural beliefs could be more complicated of an issue than for those families whose deaf child has no additional disabilities (Luterman, 2003). Parents are

likely to feel isolated because they are dealing with a unique situation and they do not fit into any specific category or group in which they could seek support or understanding. They do not have a support group that identifies the child's multiple contributing disabilities in addition to hearing loss (Carey, Crocker & Elias, 2009). The grieving process for families who are just learning that their child has a hearing loss ranges from denial to acceptance to anywhere in between (Paul & Whitelaw, 2011). The social worker working on a "special case" team must be able to think critically and outside of the box, since evidenced-based practices does not clearly exist. However, there are abundant sources that have examined different families' expression of grief and coping mechanisms for hearing loss only in their infant or child (Anagnostou, Graham & Crocker, 2007; Clark & Brueggman, 2004; Gravel & McCaughey, 2004; Jackson & Turnbull, 2004; Meadow-Orlans, Mertens, Sass-Leher, & Scott-Olson, 1997; Sjoblad, Harrison, Roush, & McWilliam, 2001) and effective counseling approaches such as parental involvement in early intervention (Ingber & Dromi, 2010; Korfmacher et al., 2008). Since no definitive counseling research has been formulated that specifically addresses dynamics, emotional issues and counseling concerning multiple disability children with hearing loss (Luterman, 2004), the social worker must have an innate set of skills coupled with diverse clinical experience for working with these families.

- Speech-Language Pathologist (SLP) - The SLP is a mandatory team member when structuring the CI team in addition to audiologists and otologists (Madell & Flexer, 2008). When arriving at cochlear implant center or clinic, the number one

benefit families are seeking are learning open-set speech perception and spoken language (Black, Hickson, Black, & Perry, 2011; Nikolopous, Dyar, Archbold, & O'Donoghue, 2005) and promoting auditory-oral, or A/O (i.e. spoken language). A/O is the typical mode of communication that receives the majority of focus in the therapy of prelingually deaf children without additional disorders (Dettman et al., 2004). Specifically, A/O programs for children with no other issues have been found to have 100% enrollment (Waltzman et al., 1997). When comparisons could be made between the outcomes in learning either A/O or total communication (e.g. cued speech, sign language etc augmentation) post-implantation/ hearing aid fitting, those who learn in an A/O environment have demonstrated better speech perception outcomes than their deaf peers using total communication (TC) modes (Geers, Archbold & Gregory, 2003; Kirk, Pisoni & Miyamoto, 2000; Waltzman, Scalchunes & Cohen, 2000). There have been fewer studies examining effects of communication mode in implanted children with *multiple* disabilities but it has revealed that choice of enrollment is extremely varied for these children (Dettman et al., 2004). Collaborating with others members such as the deaf teacher, educational audiologist and developmental pediatricians offers increasing knowledge on communication concerns, how to approach services and gaining insight on the interrelationship of behavior, cognition, social interaction, motor skills on communication outcomes and their potential growth.

In the end, the contributing expertise from each team specialist helps offer an overall analysis and growth potential in as many areas that will benefit in the development,

health and overall well-being of the child (Accardo & Shapiro, 2006). These differences of experience are important to have on a team, especially when they help to coordinate care and management of the child. Team coordination will lead to positive outcomes; however, there needs to be a precise manner in how coordination operates among team members and the family that leads to the best outcomes of care. An interdisciplinary approach underscores the key concept of integrating separate professions into what effectively functions as one big isolated unit. That is, if a family is inquiring about the candidacy evaluation process, they will be receiving a single consultation versus each specialization providing their own consultation (Paul & Whitelaw, 2011). But a true interdisciplinary approach requires more than a professional partnership that is 100% in collaboration with each other; a parent-professional cooperative partnership is the definitive solution. In other words, no interdisciplinary methods exist without parent(s) being a dynamic part of the team, offering consistent input for evaluating, planning and deciding the route of success in their child (Orchard et al., 2005).

Collectively the outcomes from differing expertise will represent their conceptual profile (King et al., 2003), allowing the team to develop specific goals for the family, which provides for a better understanding of both strengths and limitations of the child. This profile will assist the team in optimizing the amount of success their evaluation and recommendations will bring to the family for candidacy selection and offering an appropriate management strategy.

The CI team needs to help the family understand how their child's specific case will have different endpoints in achievement with this type of intervention (Accardo &

Shapiro, 2006). That is, they need to gently, yet efficiently translate the message that regardless of how much planning and management are offered, some outcomes are not realistic for certain children. Even within one distinct disability such as autism, outcomes with cochlear implants can be quite unpredictable in these children (Wiley, Meinzen-Derr, & Choo, 2008). Although there are no guarantees of outcomes for any child with a cochlear implant, a lot more variability exists within this population. It is important for all cochlear implant teams to establish a well-organized system of communication, demonstrating a fundamental obligation to act in accordance with one another and approach decisions on a common goal basis.

Based on an informal survey of pediatric cochlear implant centers, it appears that Children's Hospital of Philadelphia (CHOP) and Massachusetts Eye and Ear Institute have a highly integrated system of professionals willing to work with a family and child with multiple disabilities. Specifically, the teams are basing an appropriate cochlear implant decision on a number of outcome perspectives that is emphasizing team collaboration. Thorough and collaborative efforts for managing a child is guaranteeing that each candidacy evaluation documents the child's full potential before determining whether a cochlear implant could be the best decision for their hearing loss, particular developmental needs and family preference.

CHAPTER FOUR

Special Case Candidacy Considerations

A number of the highly renowned pediatric cochlear implant centers, including the Children's Hospital of Philadelphia (CHOP) and Massachusetts Eye and Ear Institute (Mass Eye & Ear), use an interdisciplinary approach that represents an ideal team who will assist in advocating candidacy for children with multiple disabilities. Establishing the ideal team can be difficult for many implant centers to structure; financial resources can limit the ability to employ an extensive healthcare roster and changes to the healthcare system can affect team options and advances in technology can alter how services are addressed by healthcare (Paul & Whitelaw, 2011). However, a strong effort to encourage collaborative efforts between professionals and families must be maintained to ensure the highest chances positive outcomes for the child.

Collaboration is especially necessary for interdisciplinary teams advocating development of a new approach for children with multiple disabilities cochlear implant candidacy. FDA criteria (2010) were discussed in Chapter 2, outlining standardized measures for typically developing children and suggested outcomes that teams are currently using to guide the consideration of recommending children without additional disorders. These FDA guidelines are based on the rationale that a cochlear implant will promote a typical (or at least improved) rate of speech and language development for children with a severe to profound hearing loss. That is, FDA currently states that the

major health problem of childhood hearing regards spoken language delays. Promoting adequate speech perception and spoken language development in children is warranted, as there have been years of research focusing on the negative effects that are resulting from hearing loss and nearly as many years of research on positive outcomes for addressing this issue with appropriate aural habilitation/rehabilitation, including cochlear implantation. However, addressing and managing the specific communication, learning and lifestyle needs of a child with additional disabilities in addition to severe to profound hearing loss may be significantly different from those needs of a deaf child whose development is otherwise typical. Differences in needs are not only due to the etiology of hearing loss but from other developmental areas, with those other areas contributing to functional limitations and those limitations resulting in a lower level of participation and interaction with others (Accardo & Shapiro, 2006; Tye-Murray, 2008).

Children with multiple disabilities have issues that are chronic and interactive, whose nature of etiology is difficult, if often impossible, to identify (Diefendorf et al., 2011). The relationship between delayed learning of language, communication, social interaction, and educational outcomes is complex and many of the issues may be subtle at time of referral for candidacy consideration. With hearing loss, missing underlying functional deficits are becoming more of a common concern with interdisciplinary teams. Newborn hearing screening benchmarks (Joint Committee on Infant Hearing & Screening, 2007) including for example the "1-3-6 model" indicates the strong recommendation that a child receives the screening, identification and intervention of hearing loss before those months of age, respectively. These early standards make it difficult for a team to observe any abnormal behavior, as many do not display marked

traits until the child reaches pre-school. Because of this, considerations in addition to a severe to profound hearing loss are becoming part of the cochlear implant evaluation and candidacy decision process (Edwards, 2007).

A useful approach for classifying these issues during a special case is to apply the World Health Organization (WHO, 2012) International Classification of Function, Disability and Health (ICF, 2012). This helps teams address concerns in functional ability that are clearly beyond that of degree of hearing loss and its effect on speech and language. ICF is an interdisciplinary approach focusing on issues that relate to impairments (i.e. physiological issues), activity limitations (i.e. disabilities) and participation barriers (i.e. handicaps) that are occurring in the individual child (ICF, 2012). That is, the ICF approach offers valuable information providing a broader decision analysis and observing functional strengths, and limitations in the child. Determining the etiology or specific diagnosis will aid in setting goals because it may offer information on developmental patterns, additional disabilities not yet observable in the first years of life and future prognosis of a condition (Accardo & Shapiro, 2006). Establishing a framework of candidacy in the ICF context for assessing, treating and managing children with additional disabilities or those with special health circumstances may lead to evidence-based decisions that gains official approval for implanting under these special considerations. A logical argument for implementing an ICF candidacy is that the approach is highlighting specific needs and incorporating the "big picture" on a child's condition.

This chapter expands beyond the scope of what the FDA considers appropriate for candidacy and uses an "off-label" ICF framework for making special considerations and for obtaining diagnostic information through physiological impairment, activity limitations (i.e. disability) and participations of barrier (i.e. handicap), to establish whether this type of intervention is most appropriate for the needs and outcomes for the child and their family (Tye-Murray, 2008). Physiology of *impairment is discussed first*, referring to audiologic testing and discussing circumstances in which a candidacy decision is made during the first stage of ICF approach. The process of fitting hearing aids and the trial period is second, which incorporates the candidacy process of aided hearing and language skill assessments. Special considerations for separating concomitant motor and cognitive issues are specifically addressed in order to obtain accurate information on the individual child. Thirdly, participation considerations are assessed with parent questionnaires and survey tools measuring perceived changes in the child's quality of life. Increased participation throughout the candidacy hearing aid trial allows for understanding outcome benefit on a different level of considerations for a cochlear implant decision.

Impairment (i.e. physiology)

The first step towards understanding the effects of a condition through the ICF candidacy approach is to classify the impairment (ICF, 2012). For pre-selection considerations, audiologic outcomes are collected throughout the entire cochlear implant process (Vohr, 2011), which initially includes objective measures through electrophysiology (e.g. ABR and ASSR). Otoacoustic emissions (OAEs) and

tympanometry are two additional tests that help for establishing initial type and degree of hearing loss. This unique candidacy population requires frequent audiologic monitoring through electrophysiology, including those infants who are placed in the neonatal intensive care unit (NICU). There are multiple reasons for infants requiring a stay in the NICU (Cone-Wesson et al. 2003) including newborn risk factors for hereditary hearing loss and premature birth (JCIH, 2007). Table 4-1 outlines risk factors for hearing loss that are part of the Joint Committee of Infant Hearing position statement (2007). The audiologist will be monitoring NICU patients throughout their stay at the hospital; these children are 10 times more at risk for sensorineural hearing loss (Bielecki, Horbulewicz & Wolan, 2011).

Although rare in the broader scope of special candidacy considerations, "emergency" situations can arise for a candidate when decisions for implantation must be determined as soon as possible and while the child remains in intensive care. Specifically, there may be a critical window that the team must recognize beneficial a cochlear implant will be for accessing spoken language through auditory input (Edwards et al., 2002). For example, those who have been identified with syndromes involving characteristic features of a severe to profound hearing loss, progressive deafness and/or visual impairment can make for timely decisions in promoting communication skills through listening and spoken language via cochlear implantation (Lanson, Green, Roland, Lalwani, & Waltzman, 2007; Seewald & Tharpe, 2011).

If an infant is diagnosed with a dual sensory impairment which may result from a number of situations including CHARGE or Usher syndrome, ocular pathologies and/or

progressive blindness is typically involved. The team needs to answer quickly about recommending an implant in these situations so communication options are maximized in the child's ability to learn language (Gregg, Wiork & Arvedson, 2004; Goller, 2006). The ability to use the visual system for accessing cues and communicating through manual forms of language such as American Sign Language (ASL) has become unreliable for these children because of decreased vision (Seewald & Tharpe, 2011). Edwards and colleagues (2002) examined the effect of what they termed the "audiologic window of opportunity" on speech perception development in twenty-two infants diagnosed with CHARGE syndrome. Due to a dual sensory impairment of vision and hearing loss and the urgency to promote listening, the authors demonstrated that ABR testing decreased the average age of hearing loss identification significantly; estimates on hearing loss was achievable at 4 months versus 25 months if audiologists waited to obtain hearing outcomes through behavioral audiometry (Edwards et al., 2002). Clearly waiting to identify a hearing loss will cause delays in the intervention process and success of the child. Chronic illness, multiple surgeries and medical appointments for children diagnosed with CHARGE will inevitably keep a family busy, so making a candidacy decision for a cochlear implant should be determined while the child is still in intensive care (Sampaio, Araújo & Oliveira, 2011).

If candidacy decisions are not presented as urgent, such as in the situations outlined previously, a hearing aid trial follows objective audiologic testing. Obtaining hearing loss estimates through frequency-specific air and bone conduction thresholds offer baseline information from 500 to 4000 Hz that is applied to the initial hearing aid fitting. As discussed in Chapter 2, FDA (2010) guidelines suggest that a hearing aid trial lasts for 3

to 6 months before proceeding with candidacy decisions; benchmarks were established in accordance to best practice performance level (Hyde, 2011) and gave way to the 1-3-6 rule for early hearing loss intervention (JCIH, 2007). However, this benchmark may not be realistic for children who are already dealing with concomitant developmental delays and multiple health issues. The expectation for a child to demonstrate auditory skill development or lack of progress from a 6 month hearing aid trial period may also not be realistic; it has been previously discussed within this Capstone that concomitant disabilities are already causing delays for making appropriate progress in the development of auditory skills, spoken language and overall communication (Edwards, 2007; Meinzen-Derr ,Wiley, Grether, & Choo, 2010). These slower acquisition rates are relating to other functional domains including for example cognition, physical/motor function, and. Children with additional issues should be assured a full trial year with hearing aids, which will help the team significantly assist in developing a better understanding for individual functioning levels and a child's growth potential before making a decision for cochlear implantation (Dettman et al., 2004).

In order to address the additional time that may be required for a hearing aid trial in children who are "special case", Mass Eye and Ear Institute's (2009) pediatric implant team offers free hearing aid trial services for an entire year to families of every socioeconomic status, which is known as the Pediatric Amplification Loaner (PAL) program. PAL allows every child under the care of Mass Eye and Ear to receive opportunities for gaining the full experience with hearing aids; the exchange of hearing aids for maintaining the child's hearing needs through the year is at no charge to the family. This social service exemplifies an already upstanding reputation of healthcare,

which demonstrates leadership role; other implant centers across the country should mimic implement this into special considerations for candidacy. Current and appropriate fitting practices for children with multiple disabilities are the same as for children without additional disorders (Tharpe, Fino-Szumski & Bess, 2001). The lack of distinction between these two groups of candidates calls attention to the fact that more outcome research and evidence-based decisions are needed for teams to offer better standards of care. In older children, hearing aids are to be worn throughout the candidacy period during audiologic follow up. Assessments outcomes will help for defining reasonable next steps for developing a treatment plan.

Activity Limitation (i.e. disability)

To ensure that a child with multiple issues reaches their full listening potential during a hearing aid candidacy trial, it is prudent for audiologists to obtain reliable behavioral thresholds (Madell & Flexer, 2008) through visual reinforcement audiometry (VRA) or conditioned play audiometry (CPA) testing, if possible. Providing accurate behavioral data allows for appropriate gain adjustments and hitting appropriate hearing aid targets to follow up with ABR/ASSR baseline outcomes. Despite challenges including the physical and cognitive issues that are influencing unpredictable response behavior with this group (Tharpe et al., 2006), obtaining reliable behavioral outcomes is possible for almost every child with developmental disabilities (Madell & Flexer, 2008). Specifically, Tharpe (2009) advocated that audiologists and interdisciplinary teams must "bear in mind that behavioral tests provide an indication of how an individual uses his or her hearing a very important factor when considering management needs" (p.210).

Through an ICF candidacy approach, the audiologic assessment cannot be administered to a child with multiple issues until activity limitations relating to their motor and cognitive delay are first managed with the necessary support services (ICF, 2012). Once the child is receiving the appropriate accommodations for issues unrelated to hearing-related outcomes, their performance will be reflecting the child's full potential in auditory skill and language development (Accardo & Shapiro, 2006). Environmental modifications (Accardo & Shapiro, 2006) that are made to the physical set up of the test area allowing for motor-related activity limitations to become effectively separated from auditory-related responses; nonverbal cognition tests are offering insight to the child's cognitive functioning and potential for learning language without an interfering issue of testing through verbal measures (Edwards, 2007). The following motor and cognitive-related issues are discussed in isolation in order to simplify the process of identifying underlying functional effects.

Physical and motor-related disabilities contribute to activity limitations that may interfere with gaining candidacy information on hearing aid benefit. The behavioral responses that are needed for obtaining aided VRA and CPA responses require modifying the physical set up of the soundbooth test environment (Accardo & Shapiro, 2006). Some examples of these functional accommodations include ordering customized equipment for gathering accurate pure-tone threshold responses. In particular, reliable bone conduction thresholds may not be possible to obtain for children with craniofacial disorders. The atypical size and shape of their skull and other dysmorphic physical features are preventing a secure fit and position against the temporal bone; structural abnormalities of the temporal bone may also prove difficult for maintaining appropriate

pressure against the skull. A young child with cerebral palsy may need a wooden high chair for structural support that allows for better head and neck rotation for turning towards a visual reinforcer for a correct behavioral response. An older child with CP who is able to perform CPA may require that toys are larger and to easier to grip, therefore demonstrating the appropriate conditioned response such as dropping the toy when they hear a sound. Similarly, the "drop bucket" should be placed near enough to the child that their range of extension allows the toy to fall successfully into the bucket (Madell & Flexer, 2008). All of these situations will help an interdisciplinary team separate motor or physical-related delays to adequately assess behavioral response thresholds that represents their functional strengths and limitations.

In addition to offering appropriate motor/physical services prior to testing these children, developmental screenings and assessing nonverbal cognitive levels (Edwards et al., 2009) are necessary for obtaining accurate response in children with concomitant cognitive issues. Considerations for language delay relating to severe to profound hearing loss must be separated from delays that are directly resulting from atypical developmental patterns and cognition. These two methods of assessment are bypassing cognition and developmental activity limitations, thus offering insight into the child's true potential for developing speech perception skills and learning rate for spoken language. Norms that are considering both of these factors include testing through the *Bayley Scales of Infant Development- 2nd Edition* (BSID-II; Black & Matula, 2000), *Schedule for Growing Skills- 2nd Edition* (SGS-II; Bellman, Lingnam & Aukett, 1996) and *Leiter International Performance Scale- Revised* (LIPS-R; Leiter, 2002). In particular, Leiter (2002) developed the LIPS-R test, which incorporates factors of language, cognition and motor

delay for resulting outcomes (Schum, 2000). Through hearing aid trials for children with milder forms of cognitive delay demonstrate positive changes at the level of the patient (Tye-Murray, 2008), which promotes considerations for a cochlear implant (Schum, 2004) in these children. In particular, this information will lead to better counseling of expectations for families who are considering a cochlear implant for the desired outcome of benefiting from speech and language development. Tracking aided developmental progress will help for mapping out direction for further management decisions.

Participation barriers

Difficulties that are apparent in the child do not simply exist as activity limitations that are decreasing the ability to internalize and formulate speech and language skills (Tye-Murray, 2008). Rather, these activity limitations are diminishing interactions within their external auditory environment and within home and social settings. These participation barriers lead to isolation which can be even more damaging to children with concomitant activity limitations, since these are inhibiting their ability to express their feelings of frustration. Understanding these underlying effects, special considerations of a cochlear implant allowing for increased environmental awareness and social interaction experiences may significantly improve quality of life for these children. That is, lifestyle has potential for improving through cochlear implantation, even if speech and language outcomes are not part of the intervention goals.

Throughout the candidacy hearing aid trial, parents are given questionnaires and surveys that are addressing perceived changes in simple sound awareness to communication efforts, including the *Children's Home Inventory for Listening*

Difficulties (CHILD; Anderson & Smaldino, 2000) and *Auditory Behavior in Everyday Life* (ABEL; Purdy, Farrington, Moran, Chard, & Hodgson, 2002). These two diagnostic questionnaires may help a special case team and family reflect on improved lifestyle and social and family interactions which focus on the "bigger picture" of a candidacy decision. With effective counseling by the interdisciplinary team, the ICF approach helps for offering a positive light on opting for the cochlear implant surgery based on maximizing the child's abilities to participate in social and engaging activities (Accardo & Shapiro, 2006). From a management perspective, these subjective questionnaires are not meant to generalize participation benefits for families pursuing a cochlear implant for improving quality of life. That is, benefit is not only looked at as one component such as better sound awareness. Rather, it helps create an understanding of this candidacy population so that team specialists are able to better attend to the specific needs of the child and family and for appropriately assessing outcomes from the candidacy process (Wiley et al., 2005).

In closing of assessment and special considerations for candidacy, one must truly reflect on the whole reason that families (for the most part) are pursuing intervention services for their child in the first place. Regardless of specific developmental issues or conditions, the family wants their child to have a chance at living life normally and to its fullest potential, resulting in their overall happiness. A perspective from parents whose child received a cochlear implant to enhance the quality of life is reflected here (Wiley et al., 2005):

"Special needs children go through so much that people see the cochlear implant as one more thing. I feel they need to have every opportunity available to him. I want my child to be treated like he is not special needs" (p. 796).

"So much was out of our control, if we could make one thing within our control, we need to do it" (p. 797).

CHAPTER FIVE

Postoperative outcomes

Appropriate testing and the definition of "benefit" for receiving a cochlear implant are unique to this population, as discussed in Chapter 4. The previous Chapter also suggested that interdisciplinary teams use an ICF framework for candidacy that allows for examining different options that maximize a child's potential before a decision is made cochlear implantation. Cochlear implant candidacy that recognizes the "bigger picture" is an ideal approach for considering candidacy as it incorporates all developmental issues, many of which the FDA (2010) overlooks as being important for a successful outcome. As the number of children with multiple disabilities receiving cochlear implants increases, families with truly diverse circumstances will be inquiring about outcomes and what that may mean for the developmental success of their child. Interdisciplinary teams must start framing standards for candidacy assessment, consideration and counseling, basing on levels of expectation in speech and language (e.g. activity limitation) as well as quality of life (e.g. participation). To achieve these goals, teams must work diligently to obtain postoperative information (Lee, Kim, Jeong, Kim, & Chung, 2010) which displays results for both terms of benefit (ICF, 2012). This chapter is focused on outcomes in the literature that currently describes speech and language development and changes in participation levels for these children post-implantation.

Speech Perception and Spoken Language

The rate of success in speech and spoken language outcomes for children with multiple disabilities will be different than their peers without additional cognitive, motor or sensory concerns (Edwards, Frost & Witham, 2006). Children with cognitive and/or motor delays demonstrate significantly slower outcomes for developing speech perception and spoken language, despite an age-equivalent factor to those without additional disorders (Edwards, Thomas, Rajput, 2009; Geers et al., 2003; Lee et al., 2010; Pyman et al., 2000; Trimble et al., 2008; Winter, Johnson & Vranesic, 2004). The outcome delays for speech perception development ranges from simple speech awareness through pattern perception, and for testing closed-set and open-set word recognition (Trimble et al., 2008). The postoperative speech measures such as the *Early Speech Perception* (ESP; Geers & Moog, 1990), *Category Auditory Performance* (CAP; Archbold, Lutman & Marchall, 1995), *Glendonald Auditory Screening Procedure* (GASP; Baumgartner et al., 2002), *Manchester Picture Test* (MPT; Hickson, 1987), and *Meaningful Auditory Integration Scale* (MAIS) are examples of speech perception tests that have a hierarchal rating that are capable of monitoring outcomes in children post-implantation.

Winter, Johnson and Vranesic (2004) tracked speech perception outcomes in implanted children for 3 years post-implantation. Ten children with additional disabilities and ten otherwise typical developing children who served as the control group were included in this study. Postoperative outcomes were measured by Early Speech Perception (ESP) performances, which Winter et al. (2004) described as a closed-set

word list with picture-pointing for children with limited vocabularies. The standard ESP version consists of word items that are familiar to most hearing impaired children by the age of six (Moog & Geers, 1990). A low-level ESP version has served as an alternate test measure for children less than two years old and whose vocabulary is even more limited (Black et al., 2011; Moog & Geers, 1990; Trimble et al., 2008). In the Winter et al. (2004) study, each child's progress was monitored post-implantation on an annual basis. Individual performances of the child were classified into one of four categories: detection or no pattern perception; pattern perception; some word identification; consistent word identification. Winter et al. (2004) found significant delays in word identification skills for the children with multiple disabilities compared to their age-matched peers. Specifically at one year post-implantation, eight of the ten children with multiple disabilities were unable to reach the 2nd level of pattern perception, even when they used the low-level ESP test version. In contrast, all ten children in the control group were able to achieve, at minimum, the 3rd level of "some word identification". In addition, eight children in the latter group were also able to be tested with the standard ESP version. At two years' post-implantation, the performances of four children with additional issues were able to progress to 2nd stage of pattern perception. However, only one child within the study group was ever able to achieve a 3rd level performance, which was not displayed until three years post-implantation. Dettman et al. (2004) reported a follow up study of deaf children with mild or moderate cognitive delay who demonstrated poor speech perception prior to implantation. Speech perception scores for children with mild cognitive delay improved significantly, and those for children with moderate cognitive delay improved slightly after implantation. Holt and Kirk (2005) monitored the outcomes

of 19 children with mild cognitive delay and concluded that these children benefited from cochlear implantation in terms of speech perception development.

In addition to slower development of speech perception, lower rates of spoken language have been demonstrated in implanted children with multiple and/or cognitive delays versus age-matched peers without such disorders (Meinzen-Derr et al., 2010; Nikolopoulos, Archbold, Wever, & Llyod, 2008). In the Nikolopoulos et al. (2008) study, implant teams monitored their progress in speech production, using the Speech Intelligibility Rating (SIR) (Allen, Nikolopoulos, Dyar, &O'Donoghue, 2001). Nikolopoulos et al. (2008) modified the performance rankings, since the assortment of original language samples on the test proved too difficult for children with multiple issues; many were never able to acquire the necessary spoken language skills to complete the task. Due to the increasing amount of children with multiple disabilities who have implants, the updated method was designed as a time-effective global outcome measure of speech production in real life situations (Lee et al., 2010). These real life situations have allowed children to be monitored over time post-implantation, despite cognitive or intellectual strengths associated with language (Allen et al., 2001; Nikolopoulos, Archbold & Gregory, 2005). The results of the Nikolopoulous et al. (2008) study indicated that 70% of the children with additional disabilities were able to acquire some type of "connected speech" or spoken language at the end of a 5-year post-implantation monitoring schedule. Fukuda et al. reported on the progress after implantation of one child with moderate cognitive delays. Preoperative monosyllabic speech perception scores were 0% in the auditory-only condition. Speech perception abilities continued to improve, and monosyllabic speech perception scores increased to 75% by 2 years post-

implantation. The child could produce only some voluntary voice sounds before implantation, but began to develop some spoken language 2 months after implantation. The slower progress or limited results for speech and language development is not meant to discourage teams and families from deciding on a cochlear implant for the child with multiple disabilities. Rather, it is hoped that through some of these outcome studies, teams can offer better counseling strategies during the candidacy process of these families. These varied outcomes in speech perception skills demonstrate that cognition level is an important factor that should be stressed during candidacy. Rates of progress and overall benefit in speech and language will also depend highly on the frequency and type of intensive therapy as well as education and family support. Building on the current data will lead for solid framing of expectations and counseling strategies for families.

Participation

Although speech and language skills may not develop the way some families hope they will once their child receives a cochlear implant, the majority of children do demonstrate increased levels of participation and improved quality of life (Tye-Murray, 2008; Wiley et al., 2005), which may be the outcome of benefit that many parents are seeking for their child (Edwards et al., 2006). Participation outcomes, which are mainly a result from small case studies, have reported improvements including more environmental awareness (Waltzman, Scalchunes & Cohen, 2000), behavioral compliance (Bass-Ringdahl et al., 2010), social play (Edwards, 2007) and attempts in communicating with others (Brady & Bashinski, 2008).

Brady and Bashinski (2008) suggested that early communication programs may be maximally effective when children with severe disabilities are taught to combine technology (e.g. cochlear implant) with unaided communication responses including natural gestures and non-speech vocalizations. The authors examined 9 children who were classified as "deaf-blind", although hearing loss ranged from mild to moderate degrees in 4 participants. The remaining 5 children presented with bilateral severe to profound deafness and who wore a cochlear implant on one ear and were unaided in the contralateral ear. Due to the complex nature of disabilities including cognitive delay a non-standardized Wisconsin Behavior Rating Scale (WBRS; Song et al., 1980) test was given to each child. Throughout "several months", the children learned communication through intensive therapy known as pre-linguistic milieu teaching (PMT; Yoder & Warren, 1998), which taught them to communicate with gestures and vocalizations. Brady and Bashinski (2008) reported that each child initiated more attempts to communicate with the speech language therapist and family after enduring this habilitation program. In accordance with these efforts, all participants had met at least one goal in their individual education plan (IDEA, 2004) that targeted an aspect of symbolic communication. Perceived benefits were noted to significantly improve in a Wiley et al. (2008) study, which examined 20 children with a variety of additional issues. In summary, these studies that documented improvements in activity level and participation restrictions allows for the argument that all children are capable of benefiting from a cochlear implant despite the severity of concomitant disabilities.

CHAPTER SIX

Conclusion

Families of children with multiple disabilities who are looking for different intervention opportunities are typically doing so because they hope to give their child, in the end, a better chance at experiencing growth in lifestyle, development and happiness. In order for healthcare and intervention services to approach this type of intervention, professionals must establish an interdisciplinary team to work with this population. Team collaboration is mandatory for a successful system of management and an interdisciplinary approach reinforces family-directed care, which is a reminder that parents are always the "bottom line of assessment, treatment and management and that planning is to be formulated specific to the child's needs and family preference. A cochlear implant team must offer a thorough candidacy assessment through the lens of the World Health Organization International Classification and Function (ICF) which assesses and considers those functional domains that the team decides to be most important for managing and allowing the individual child to reach their full growth potential. Accessing sound, in this case via cochlear implantation, does not restrict growth potential to a relatively simplistic goal of speech and language; hearing is clearly more important to individual well-being than just for communicating through spoken language. Having adequate access to sound helps for improving a plethora of developmental factors and personal achievements for which a child is capable of

obtaining throughout their life. A child with multiple disabilities is capable of achieving benefit and success through a cochlear implant, and whether those outcomes equate to benefits in communication or to increase in participation for overall improved quality of life does not dissuade from the fact that this population is just as deserving to all the rights of implanting a child without additional issues.

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